Coexistence of Granuloma Annulare and Castleman’s Disease: A Case Report

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INTRODUCTION

Granuloma annulare (GA) is a self-limited disorder that may present as localized or general. Localized GA, the most common form, presents as an erythematous annular plaque without scale. The generalized form of GA presents as widespread erythematous papules and plaques over the trunk and extremities. A cross-sectional study in the United States found a female to male prevalence ratio of 3:1 with a higher prevalence in the fifth decade of life.¹ The etiology of GA is largely unknown; however, a number of potential inciting factors have been reported including viral infections, drug exposure, trauma, and insect bites.²

Castleman’s Disease is a rare heterogenous lymphoproliferative disorder that encompasses a spectrum of clinicopathologic manifestations. Unicentric disease affects one lymph node or chain of lymph nodes with primarily hyaline vascular changes. The multicentric form affects multiple chains of lymph nodes with primarily plasmacytic changes. The pathogenesis is related to hypercytokinemia due to dysregulated interleukin-6 (IL-6) activity in unicentric disease and dysregulated IL-6 and human herpes virus-8 in multicentric disease.³

We report a case of concomitant granuloma annulare and Castleman’s Disease.

CASE REPORT

We present the case of a 58-year-old male who presented with a worsening, diffuse rash for several months. On physical exam, the rash consisted of erythematous papules coalescing into plaques on bilateral upper and lower extremities, posterior neck, and trunk with associated pruritis (Figure 1, 2). Biopsy of the right forearm showed perivascular and interstitial mononuclear infiltrate with focal palisading granulomas containing central necrobiosis with mucin deposition, consistent with GA. The associated rash became more generalized to include his trunk and extremities and he was diagnosed with generalized GA. This patient presented two years later with fatigue, night sweats, and widespread lymphadenopathy. He subsequently underwent CT and lymph nodes biopsy and was ultimately diagnosed with multicentric Castleman’s Disease.

DISCUSSION

The simultaneous presentation of granuloma annulare (GA) and Castleman’s Disease in our patient raises questions about possible shared pathogenic pathways. Castleman’s...
Figure 1. Erythematous papules coalescing into plaques on bilateral upper extremities, posterior neck, and back.

Figure 2. Erythematous papules coalescing into plaques on bilateral lower extremities.
Disease hinges on hypercytokinemia, specifically due to dysregulated IL-6. In contrast, while the exact mechanisms underlying GA remain elusive, recent studies have emphasized its inflammatory basis, showcasing the involvement of Th1/innate cytokines, Th2 pathways, and notably, the JAK-STAT signaling pathway.\(^4\)

Given that the JAK-STAT pathway is pivotal for cytokine responses, including IL-6, one might speculate a possible overlap in cytokine dysregulation between these two conditions. This convergence might provide insights into the potential shared etiological factors or triggering events.

Furthermore, while external factors such as viral infections, trauma, or drug exposures have been proposed as potential triggers for GA, it remains to be explored if any of these could also influence the onset or progression of Castleman’s Disease.

This unique coexistence of GA and Castleman’s Disease in our patient underscores the potential intertwined nature of their pathogenesis and the importance of investigating deeper into their interrelated inflammatory mechanisms.

**Conflict of Interest Disclosures:** None

**Funding:** None

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